

Behçet's disease-related superior vena cava syndrome and bleeding downhill varices: A rare complication

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ABSTRACT

Obstruction of the superior vena cava (SVC) due to any cause results in development of venous collaterals in the upper part of the esophagus, known as “downhill” varices. Although rare, bleeding can be life-threatening. Presently described is case of Behçet's disease-related SVC occlusion in a patient who presented with gastrointestinal bleeding from upper esophageal varices.

Keywords: Behçet's disease; downhill varices; superior vena cava syndrome.

INTRODUCTION

The most common esophageal varices are located in the distal part of the esophagus and are almost always associated with portal hypertension. This type is called “uphill,” due to upward direction of blood flow. Upper esophageal varices are less common than distal uphill type, and referred to as “downhill” varices (DEV). These varices are mostly associated with superior vena cava (SVC) obstruction secondary to mass effects of tumor or venous thrombosis. Increased blood flow into esophageal veins has been considered etiology of DEV in rare cases.

Presently described is case of bleeding from downhill varices caused by SVC occlusion in a patient with history of Behçet's disease.

CASE REPORT

A 40-year-old male was admitted to hospital with sudden-onset hematemesis. He seemed pale and vital signs revealed

slightly low blood pressure (100/70 mmHg blood pressure with heart rate of 92 bpm). Rectal examination revealed solid black stool, but no other abnormalities were present on physical examination. Laboratory values on admission showed anemia (9.0 g/dL hemoglobin (normal: 12.5–16 g/dL) and 29.8% hematocrit (normal: 37–47%) with normal platelet count and coagulation parameters. Medical history included diagnosis of Behçet's disease in 1995. In 1996, the patient had intracranial hemorrhage due to prolonged prothrombin time as result of uncontrolled use of Coumadin (Bristol-Myers Squibb Co., New York, NY, USA). After recovery, Coumadin was exchanged for acetylsalicylic acid. Treatment regimen since then had been immunosuppressive (azathioprine 100 mg/day) and antiaggregant (acetylsalicylic acid 100 mg/day). After transfusion of a unit of red blood cells and attaining hemodynamic stabilization, esophagogastroduodenoscopy was performed with initial diagnosis of peptic ulcer bleeding due to acetylsalicylic acid use. Endoscopic examination revealed grade II esophageal varices in upper part of the esophagus, although neither active bleeding nor stigmata of recent bleeding were noted (Fig. 1). Distal esophagus, stomach, and duodenum were entirely normal and no peptic ulcer was detected. Most likely diagnosis was bleeding from downhill varices. As there was no active bleeding, the patient was managed conservatively. Contrast-enhanced computed tomography scan of the chest was performed to determine etiology of varices and it showed marked stenosis of the SVC and collateral veins on the chest wall (Fig. 2). The patient's general condition and hemoglobin levels remained stable and he was discharged a few days later.

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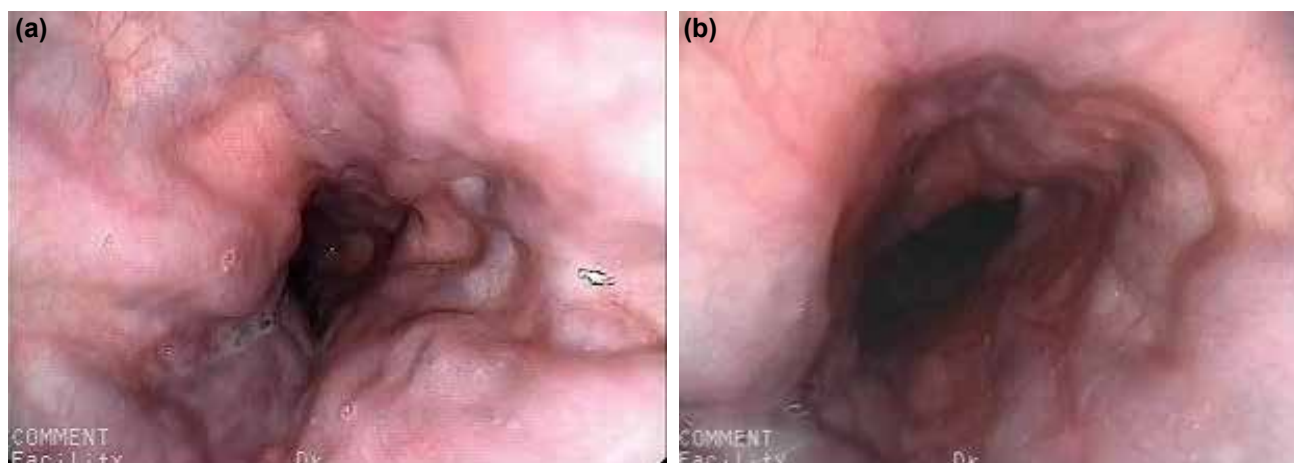


Figure 1. (a, b) Esophagogastroduodenoscopy revealing “downhill” varices in the upper part of the esophagus.

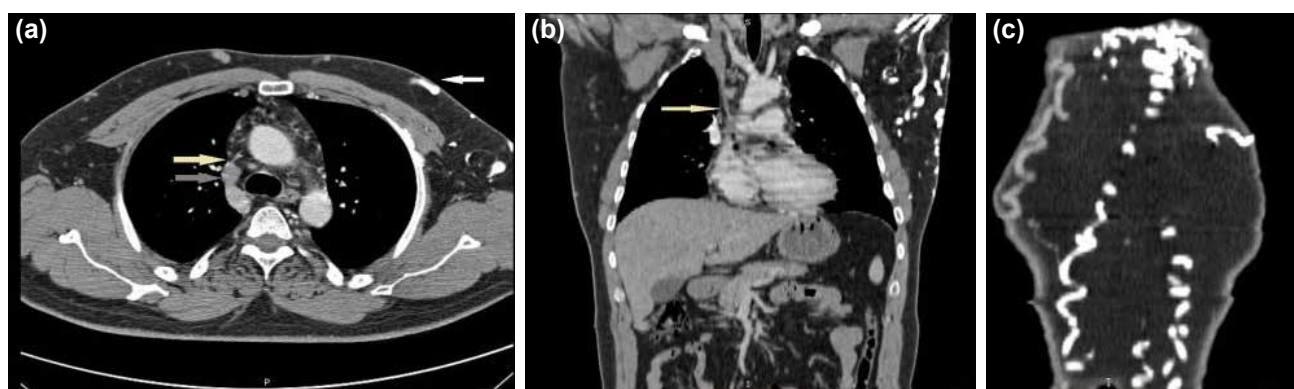


Figure 2. Contrast-enhanced computed tomography of the chest. (a) The obvious occlusion of the superior vena cava (SVC) (yellow arrow), with dilated azygos vein (grey arrow), and collateral veins on the chest wall (white arrow). (b) Coronal reformatted images indicating tight SVC occlusion (arrow) and (c) subcutaneous collateral veins on the chest and abdominal walls.

DISCUSSION

Behçet's disease (BD) is a chronic multisystemic vasculitis of unknown origin, first described by Turkish dermatologist Hulusi Behçet in 1937.^[1] It is characterized by recurrent oral and genital ulcerations, as well as inflammation of the eyes. Many other mucocutaneous, musculoskeletal, cardiovascular, gastrointestinal, and neurological systems can be affected. Vasculitis affects both arterial and venous systems, and involves all sizes of vessels, although arteritis is less frequent.^[2] Involvement of great veins, such as SVC, is well-defined but rare complication, occurring in less than 2% of patients with BD.^[3] When SVC obstruction occurs, blood from the head, neck, and upper extremities cannot directly reach the right atrium. One of the venous collaterals through which blood flows to bypass the obstruction is deep esophageal veins, resulting in proximal esophageal varices.^[4] Due to retrograde blood flow, they are also known as DEV. This type of varices is rare, and has much lower risk of bleeding than classic (uphill) distal varices (9%). DEV are mostly due to SVC syndrome, which may be related to various etiological factors. Mass effect of malignancy is predominant (95%); DEV secondary to SVC in a patient with BD is very rare. Pubmed

search revealed only 7 reported cases. As the data are so limited, there is no consensus on treatment of such DEV; however, underlying cause of obstruction should be treated. As in many forms of vasculitis, immunosuppressive agents are primary therapy in BD with major venous involvement. Anticoagulant or antiaggregant therapy is controversial, as thrombus is adhered to vessel wall and embolism is rare.^[5] However, these therapies may facilitate bleeding in case of varices. This case demonstrates that DEV could be possible etiology of gastrointestinal bleeding in a patient with BD; therefore, upper esophagus should be evaluated carefully in such patients.

Conflict of interest: None declared.

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OLGU SUNUMU - ÖZET

Behçet hastalığı ile ilişkili süperior vena kava sendromu ve kanayan downhill varisler: Nadir bir komplikasyon

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Süperior vena kava'nın herhangi bir nedenle tıkanması, yemek borusunun üst kısmında venöz genişlemeler ile sonuçlanır ve bunlar "downhill-aşağı yönlü" varisler olarak adlandırılır. Nadir olmasına rağmen, kanamaları hayatı tehdit edici olabilir. Bu yazıda, Behçet hastalığı'na bağlı süperior vena kava tıkanıklığı sonucu gelişen ve kanayan üst yemek borusu varisli bir olgu sunuldu.

Anahtar sözcükler: Behçet hastalığı; downhill varisler; süperior vena kava sendromu.

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